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Case report and review of the literature

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Dear Editor,

my co-authors and me would like to submit the attached Case Report for reviewing and publication in Clinical Neurology and Neurosurgery. Thank you very much.

There are no Conflicts of Interest and Ethical Adherence. There is no financial disclosure.

Kind regards, David Bellut

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Keywords:

Cystic lesion, clivus, CSF, rhinorrhea, biopsy

Introduction:

Lesions of the clivus include meningiomas, chordomas, metastatic lesions, chondrosarkomas, teratoma and pituitary adenomas. Cystic lesions of the clivus however are mainly chordomas or immature teratomas. Only some cases of meningoceles, metastases and pituitary adenomas presenting as cystic lesions of the clivus have been described[1,4]. We discuss the case of an extended CSF containing cystic lesion of the clivus.

Case report:

A 65 year presented with blackout, previous vertigo and headache for which her family doctor ordered an MRI scan. A cystic lesion in the clivus was detected incidentally. 12 years prior she had a carcinoma of the larynx that was treated by surgery and radiotherapy (72 G; the clivus was not in the field of radiation).

At time of admission the patient complained about minor headache but no other neurological symptoms could be found. MRI showed an extended cystic T1-hypo-, T2-hyperintense homogenous lesion of the clivus with extension to the pterygopalatine fossa and the right optic nerve canal (Figure 1-3). There was no connection to the CSF system visible and there was no pathological contrast enhancement. Signs of vascular leucencephalopathy and a small development venous anomaly (DVA) within the left caput nuclei caudatus were found but no other secondary intracranial pathology. The preoperative computed tomography (CT) scan showed massive erosion of the bony structures of the clivus and the pterygoid (Figure 1). The irregular bony delineations and the heterogenic presentation on

imaging of tumors of the clivus in particular of chordoma, the latter was suspected and a biopsy was performed.

Surgery was performed with intraoperative navigation using a transnasal endoscopic approach through the right nostril. After a sphenoidotomy the mucosa was removed. Elevating the mucosa a mild loss of water clear fluid was identified and sampled for diagnostics. Opening the anterior thin bony membrane of the cystic lesion, lead to loss of large amount of water clear fluid, highly suggestive for CSF. The wall of the cystic lesion could be partly dissected and was sent for histopathological examination. Macroscopically the cystic lesion had an arachnoidal cyst like appearance. Exploration of the cyst showed that there was neither bony covering of the right optic nerve nor the cavernous segment of the right carotid artery. Closure of the operative opening was achieved with dura substitute and fibrin glue. Postoperative imaging with a CT scan showed partial removal of the cystic lesion and intracranial collection of air in the area of the basal cisterns, the ventricular system and the frontal subdural space.

The cytological diagnostic showed CSF with a cell count of 35 mononuclear cells and 1000 erythrocytes. The histopathological diagnostic showed the cyst wall as membranous structure covered with respiratory epithelium with subepithelial fibrosis and without sero-mucous glands. These findings were in accordance with the diagnosis of a CSF containing membranous cystic lesion of the clivus.

The patient could be discharged on the fourth postoperative day without signs of neurological symptoms or CSF rhinorrhea. 10 days after surgery the patient was readmitted because of an episode with blackout, headache, subconsciousness and CSF rhinorrhea. The CT imaging showed a massive pneumocephalus. On surgery, a CSF fistula could be detected posterior to the second geniculum of the right internal carotid artery. This fistula could be closed with abdominal fat, fascia lata and fibrin

glue. A lumbal drain was placed and 5ml/h CSF was drained for 8 days. The patient could then be discharged on the 13th postoperative day without signs of neurological deficits, rhinorrhea or hydrocephalus.

Three months and one year after second surgery the patient was seen for follow-up and reported no signs or symptoms of CSF rhinorrhea, was neurologically healthy and the CT scan showed complete removal of the cystic lesion and no pathological contrast enhancement.

Discussion:

CSF containing cystic lesions of the bony skull are extremely rare. They have mostly been reported after head trauma with skull fracture as intradiploic arachnoid or leptomeningeal cysts[5]. There is one case with appearance of a CSF pseudocyst as a rare complication after ventriculoperitoneal shunt malfunction. In most reported cases bony cysts are located in the occipital bone[5]. The pathomechanism of the cyst growth after trauma has been discussed widely in the literature. Most authors explain the increase of the cyst as an arachnoidal cyst gaining size due to the pressure of the pulsating CSF at the site of the fracture and consecutive dural opening[5].

In the present case the membrane of the cystic lesion found during the first surgery for biopsy was extremely thin, so that a spontaneous CSF rhinorrhea could have occurred without treatment. However to our knowledge there are only few cases published in the English literature with spontaneous transclival CSF rhinorrhea[1,3]. Focal atrophy of the bone, congenital bone defects and developmental defects in the sphenoid bone in addition to elevated intracranial pressure being responsible for the rhinorrhea were discussed as there was a small fenestration of the bone of the clivus found in one case. However there were no signs or history of elevated intracranial pressure found in most of the cases[3]. In comparison to those few previously reported

cases, our patient presented with massive erosion but no discontinuity of the outline of the clival bone.

Another often discussed reason for spontaneous CSF rhinorrhea is Sternberg's canal which is the lateral craniopharyngeal canal. A patent canal has been found in nearly 4% of all human adults[2]. The published cases of CSF rhinorrhea due to the presence of Sternberg's canal are usually connected with sphenoid sinus CSF leaks and not with transclival CSF leaks[2]. Furthermore the existing of a canal lateral to the originally described Sternberg canal seems to have a much higher incidence of spontaneous CSF rhinorrhea[2]. There are cases of meningoceles through Sternberg's canal leading to bony destruction of the pterygoid base and the sphenoid sinus wall but no involvement of the clivus as in our patient has been described.

To our knowledge no cases of CSF containing cysts of the clivus have been reported in the literature. Moreover, the bony dehiscence including a fistula at the foramen lacerum (or second geniculum of the ICA) as detected on surgery in our case seems to be very unusual. As there are neither signs of posttraumatic changes in the preoperative imaging nor any head trauma or cranial surgery in the medical history of the reported patient the pathomechanism for the development of the cystic lesion in this patient remains unclear.

We did not find any comparable case in the literature. The present case of a patient with an intraclival CSF containing cyst is extremely rare and was misdiagnosed as a chordoma. Biopsy was performed to confirm diagnosis despite the location of the pathology with predictable high risk of CSF rhinorrhea. Pathological diagnosis was established but the patient developed complications and had to be readmitted to the hospital for further surgical treatment. Endonasal and transsphenoidal endoscopic closure helped to seal the CSF fistula. The patient could be discharged and showed no deficit or complications at follow-up.

Conclusion:

The present case of a patient with a clival CSF containing cystic lesion seems to be an unique case as there is no similar description in the literature. The lesion was misdiagnosed as a chordoma and biopsy was performed despite predictable high risk of complications. A well delineated lesion in the clivus without signs of infiltration of adjacent structures showing homogenic pattern on MRI rather should be followed with imaging than sent for biopsy. Particular attention should be paid if such a tumorlike lesion is isodense on MRI with CSF. However, if imaging proof rapid growth or signs of malignancy, surgical biopsy remains the modality of choice for diagnosis and treatment.

Figure legends:

Figure 01: Preoperative CT scan, sagittal view

Figure 02: Preoperative CT scan, sagittal view

Figure 03: Preoperative MRI study, sagittal view

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Figure 01

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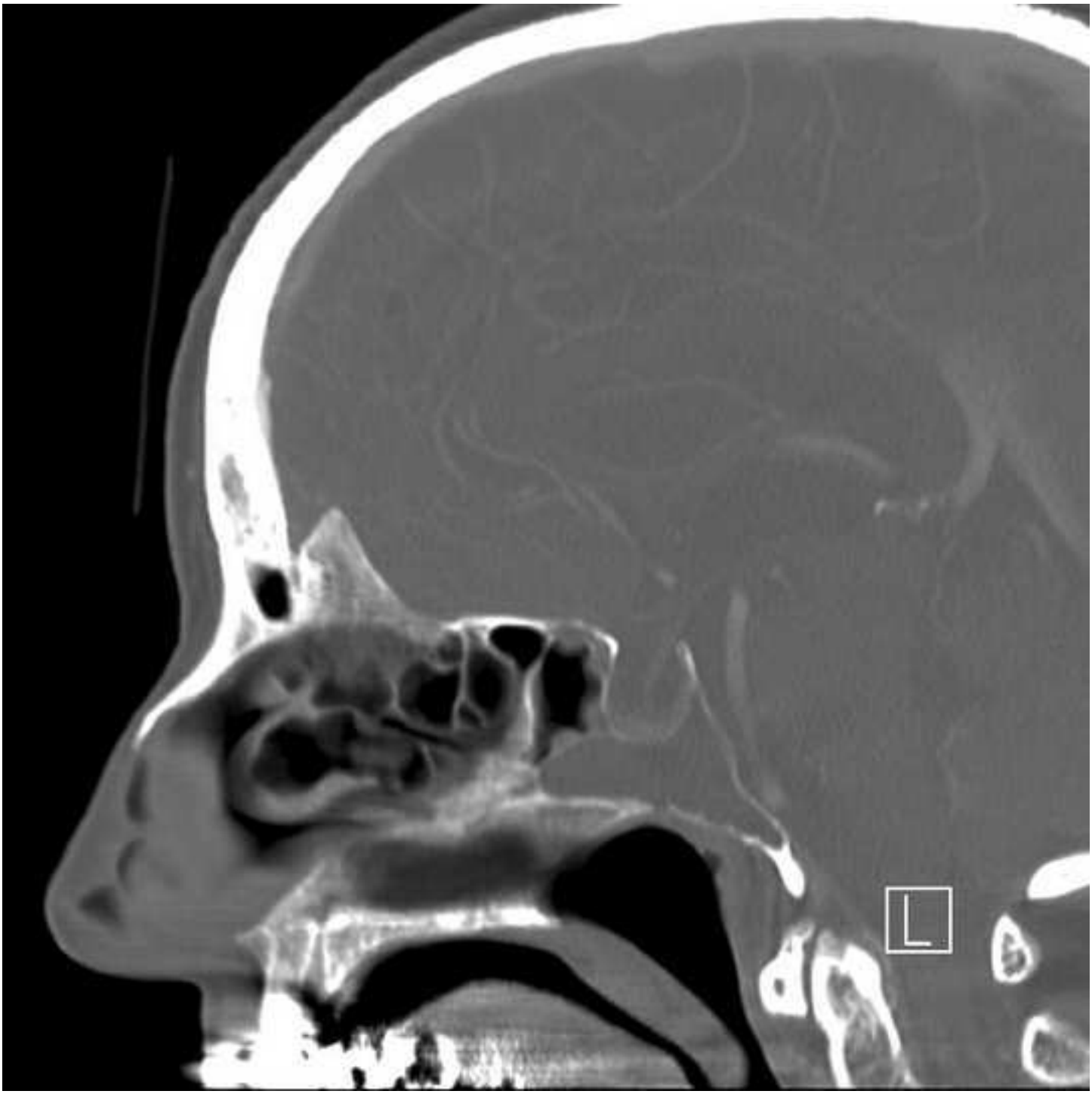

Figure 02

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Figure 03

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